312 Pre-emptive transjugular intrahepatic porto-systemic shunt (TIPSS) for portal hypertension from cystic fibrosis liver disease: 1 year evaluation

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Background: Though oesophageal variceal bleeding is regarded as the main complication of portal hypertension (PH) from cystic fibrosis (CF) liver disease increasing splenomegaly causes earlier concern when provoking abdominal discomfort, appetite loss by gastric compression and hypersplenisme. Treatment lacking, the potential of preventive TIPSS, before variceal bleeding, was investigated.
Patients: In 5 children (M4.9 y; range 4–7.5 y) annual ultrasound, fibroscan and systematic clinical examination detected liver abnormalities. When they persisted TIPSS was installed after 1.3–4.5 y (M2.5 y) to arrest progression of PH.
Results: In the first year TIPSS did not change PH complications i.e. existing abnormalities remained while non present ones dit not develop. It slowed progression of PH consequences on spleen size and function but did not reverse existing ones. No oesophageal bleeding, ascites or side effects occured. Subjective complaints and quality of life improved. In all patients TIPSS placement caused a persistent deterioration of serum albumin and INR, remaining unaltered since.

	Patient				
	1	2	3	4	5
Clinical splenic enlargement*	+++/+++	+++/+++	+/+	0/0	0/0
Platelets (10^3/μL)	55/63	62/38	264/269	242/197	196/233
INR *	1.34/1.61	1.34/1.80	1.03/1.1	1.00/1.20	1.01/1.13
Albumin (g/dL)*	3.94/3.66	3.31/3.7	4.46/4.22	4.33/4.1	4.21/3.7

*At TIPSS placement/1 y later.

Conclusion: In the first year after placement TIPSS arrested progression of PH and its consequences on spleen and oesophageal varices. If confirmed pre-emptive TIPSS can be promising if timed early, at the first signs of progressive LF, before splenic enlargement and oesophageal varices are established.

313 A single center experience of liver transplantation in CF patients

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CF related liver disease (CFLD) is a significant cause of morbidity and mortality. Portal hypertension increases the risk of death and LTx can be the only treament if lung function is only moderately impaired.
Aim: Retrospective review of cases considered for LTx over the last 9 years.
Methods: From 1990 to 2009 380 pts received 429 LTx, out of these 7 with CF (1.63%) since 2001. Before LTx all CF pts presented liver cirrhosis, portal hypertension and oesophageal varices bleeds. Pt 5 underwent splenectomy and had chronic HCV infection. Pt 4 was qualified as an urgent recipient due to acute liver failure.
Results: 7 CF pts received 8 allografts, 2 from related (pts 1, 3) and 6 from cadaveric donors.

Patients' data pre and post LTx

Pt	Gender	CFTR mutations	Waiting time (mo)	age at LTx (yr)	FEV1 pre/post (%pred)	BMI pre/post	Survival time (yr)
1	M	dF508/dF508	6	8.3	91.46/84.68	14.6/15.7	8.7
2	M	dF508/G542X	12	7.6	103.98/89.17	14.6/17.4	7.5
3	F	dF508/dF508	6	10	81.71/–	16.0/–	0.2
4	F	dF508/R553X	32	13.8	62.08/54.38	17.1/16.6	5.5
5	M	dF508/dF508	1	18	102.95/79.55	18.8/19.7	3.8
6	M	dF508/R437P	2	7.3	130.60/112.20	14.0/13.8	0.3
7	M	dF508/dF508	6	12.2	80.08/–	17.1/15.9	0.2
			mean±SD 9.29±10.63	mean±SD 11±3.9			mean±SD 3.7±3.6

The following major complications occured: thrombosis of hepatic artery (pts 3, 4), acute rejection (pts 3, 5), renal insufficiency (pt 5), biliary complications (pts 1, 2, 7), respiratory infections (pts 1, 3, 4), diabetes (pts 1, 4). Pt 3 who underwent reLTx for primary allograft rejection died of systemic multiorgan failure.
Conclusions: Respiratory status has remained unchanged except pt 4. There was no or slight improvement in nutritional status. LTx is acceptable treatment for pts with CFLD and mild to moderate pulmonary disease.

314* Long term outcome of liver transplantation (LT) for patients with cystic fibrosis liver disease (CFLD)

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Aim: To evaluate long term outcome of LT in adults (A) and children (C) with CFLD.
Methods: Retrospective review of adults (21, 16M/5F) and children (19, 11M/8F) with CFLD who underwent LT in Birmingham between 1987–2009. Data included demographics, indications, nutritional data [BMI (A), height/weight z-scores (C)], lung function [%FEV1/FVC (A), FEV1/FVC z-scores (C)], renal function (calculated GFR) and post LT complications.
Results: 1and 5 yr actuarial survival rates were 85% and 64% for (A) and 90% and 85% for (C) respectively, comparable to survival rates for other indications.

	at LT	6 mths	12 mths	24 mths	60 mths
median %FEV1	49.2	49.5	48.8	41.3	37.6
FEV1 z-score	−1.51	−1.07	−1.00	−1.22	
BMI	19.7	19.3	19.2	19	19.6
Wt-z-score	−0.74	−1.15	−1.27	−0.99	−1.72
Ht z-score	−1.25	−1.59	−1.68	−1.75	−1.35
median cGFR	108.7	99.7	81.7	72.5	84.3
median cGFR	120.9	64	70.9	77.1	83.1

Lung function (LF) stabilised after LT up to 48(A)/24(C) mths, then deteriorated as per non-LT CF patients. Admissions for chest sepsis reduced in both groups. Patients (A) with FEV1 40–50% predicted had similar outcome to those with FEV1 > 50%. Late deaths in each group were from respiratory complications. LT did not improve nutritional status, and pre-OLT BMI did not alter 1 or 5 yr survival. cGFR trends were comparable to non-CF LT patients.
Conclusion: LT is effective palliative treatment for CFLD, stabilises respiratory disease but does not improve nutrition.

315 Current practices in the diagnosis and management of cystic fibrosis related diabetes (CFRD) and impaired glucose tolerance (IGT) in CF centres in the UK

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Increasing life expectancy has resulted in more people with CF developing abnormalities of glucose metabolism. The CF Trust published recommendations for screening and management of CFRD in 2004.
Methods: In 2008 questionnaires were sent to 50 adult (A) and paediatric (P) CF centres. Data regarding current screening and management practices were determined and compared to the guidelines.
Results: 40 (80%) questionnaires were returned (20A, 20P). In P oral glucose tolerance tests (OGTT) were commenced at 10, 12 and 13 yrs in 11, 5 and 1 centres respectively. The majority of centres performed annual OGTT (15A, 16P). To screen for CFRD 2A centres used HbA1c, 1A and 2P blood glucose monitoring and 2P used high random glucose as a trigger to initiate an OGTT. Patients identified as having IGT were monitored in a variety of ways e.g. repeat OGTT in 6 months, blood glucose profile/continuous glucose monitoring, assessment of symptoms and clinical status. 5A and 5P centres would consider treatment with insulin for IGT. Following a diabetic OGTT a period of home blood glucose monitoring was initiated at 14A and 4P centres and treatment commenced dependent on the results. P centres were more likely to refer to the Diabetic Team for management decisions and more likely to commence insulin therapy without blood glucose monitoring. 16A and 6P centres had joint Diabetic/CF Clinic facilities. Details of dietary and medical management will be reported.
Conclusion: Despite publication of CFRD guidelines four years prior to the survey there remains wide variation in practice in UK CF Centres. In light of more recent evidence and availability of technology guidelines should be reviewed.